Journal of Surgical Oncology 2000;73:174–178

Congenital Pleuroperitoneal Communication in a Patient with Pseudomyxoma Peritonei

SOPHIE R. PESTIEAU, MD,[1] RICHARD WOLK, MD,[2] AND PAUL H. SUGARBAKER, MD, FACS[1]*
[1]The Washington Cancer Institute, Washington Hospital Center, Washington, DC
[2]Kaiser Permanente Medical Offices, Fresno, California

Background and Objectives: Pseudomyxoma peritonei syndrome is a rare disease arising from a perforated appendiceal adenoma. The syndrome is characterized by progressive accumulation of mucinous ascites and tumor within the peritoneal cavity. Direct extension of pseudomyxoma peritonei to the pleural cavity is uncommon and has been associated with surgical penetration of the diaphragm at the time of cytoreduction.
Methods: We review the case of a patient who presented with mucoid peritoneal and pleural fluid consistent with spontaneous pleural spread of pseudomyxoma peritonei.
Results: Surgical exploration confirmed direct pleuroperitoneal communication by macroscopic diaphragmatic fenestration.
Conclusions: This is a rare phenomenon. We outline a therapeutic approach to be applied when pleural involvement is suspected in patients with pseudomyxoma peritonei syndrome.
J. Surg. Oncol. 2000;73:174–178.

KEY WORDS: **diaphragmatic fenestration; intraperitoneal chemotherapy; intrapleural chemotherapy**

INTRODUCTION

Pseudomyxoma peritonei syndrome is a rare disease characterized by progressive accumulation of mucinous tumor and ascites throughout the abdomen and pelvis [1,2]. The primary tumor is an adenoma of the appendix that perforates and spreads mucus-producing epithelial cells throughout the abdomen that eventually fill the peritoneal cavity. Although this tumor does not invade nor metastasize, it becomes fatal in the absence of special treatments. The pathophysiology of pseudomyxoma peritonei syndrome is characterized by tumor distribution that follows the flow and resorption of peritoneal fluid, resulting in massive accumulation in the greater omentum and undersurface of the diaphragms [3,4].

Extra abdominal spread of pseudomyxoma peritonei is a rare occurrence. The medical literature provides only a few reports of pleural dissemination in the pseudomyxoma peritonei syndrome [5,6]. The patient reported here presented with pleural extension of a perforated appendiceal adenoma that occurred as a result of three diaphragmatic fenestrations. This study focuses on a rare phenomenon and outlines a therapeutic approach to apply when pleural involvement is highly suspected or confirmed at time of surgery.

CASE PRESENTATION

In September 1998, a 52-year-old white female underwent surgical exploration at another institution for repeated episodes of menorrhagia. At the time of operation, a large amount of gelatinous material was found within the pelvis and right paracolic sulcus. The mucus was removed. In addition, a hysterectomy, bisalpingo-oophorectomy, and appendectomy were performed. Pathological examination revealed a mucinous cystadenoma of the appendix. The patient was given the diagnosis of pseudomyxoma peritonei syndrome. Her postoperative course was uneventful. In January 1999, follow-up computed tomographic (CT) scans of the

*Correspondence to: Paul H. Sugarbaker, MD, FACS, The Washington Cancer Institute, Washington Hospital Center, 110 Irving Street, NW, Washington, DC 20010. Fax: (202) 877-8602.
E-mail: sap6@mhg.edu
Accepted 11 November 1999

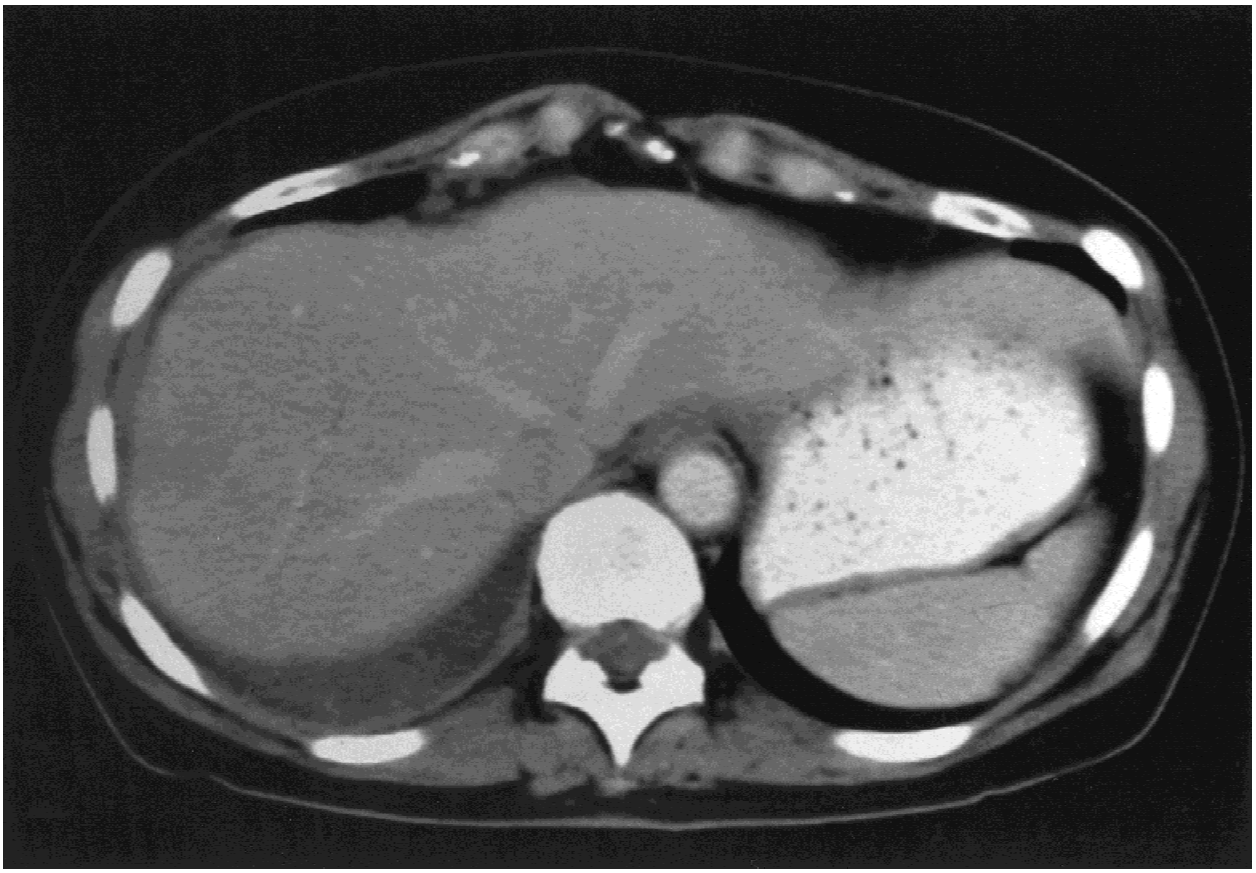

Fig. 1. CT showing mucinous tumor surrounding the liver.

chest, abdomen, and pelvis revealed perihepatic (Fig. 1) and pelvic recurrence. There was also a right pleural effusion characterized by nodular areas, suggesting mucinous tumor accumulation (Fig. 2). Because of recurrent disease in her abdomen with possible pleural spread, the patient was referred to our institution for definitive therapy in February 1999.

Upon surgical exploration, tumor nodules were found in the greater omentum, on the surface of the spleen and liver, and on the right pelvic side wall. There was also a 2-cm thick accumulation of mucoid material beneath the right hemidiaphragm. As the mucinous tumor was debrided from the undersurface of the right hemidiaphragm, three fenestra were visualized in the central tendon of the hemidiaphragm. The three defects were 3 mm in diameter, circular, and probe patent (Fig. 3). The central portion of the diaphragm was excised. Upon entering the chest through the diaphragm, there was 400 cm^3 of free mucus in the right pleural space. Of significant interest were separate bullous mucoid accumulations intimately associated with each fenestra (Fig. 4).

Once the peritoneal and the right pleural cavities were free of tumor, heated intraoperative intraperitoneal chemotherapy (10 mg/m^2 of mitomycin C in 3 L of 1.5% dextrose peritoneal dialysis solution at 43°C) was administered. The entire peritoneal cavity and the right pleural space were perfused for 90 min using a Tenckhoff catheter and four suction drains. These were removed at the end of the perfusion. She tolerated the procedure well. Her postoperative period was unremarkable. Pathological examination of the resected specimens was compatible with disseminated peritoneal adenomucinosis [7]. At 5 months postcytoreduction, the patient is currently without clinical evidence of disease. No other therapy has been recommended and the patient will be followed by CT of the chest, abdomen, and pelvis on a 6-month basis.

DISCUSSION

Until recently, treatment of pseudomyxoma peritonei syndrome was only directed toward palliation; surgery was performed to debulk mucinous ascites and tumor in order to delay the inevitable fatal outcome. Better understanding of the natural history of the disease and its distribution to peritoneal surfaces has led to an important change in the therapeutic approach, and therefore in the outcome. Introduction of aggressive cytoreduction with peritonectomy, combined with heated intraoperative intraperitoneal chemotherapy, has led to a large percentage of long-term survivors. The 5-year survival rate with the combined treatment approaches 85% [1].

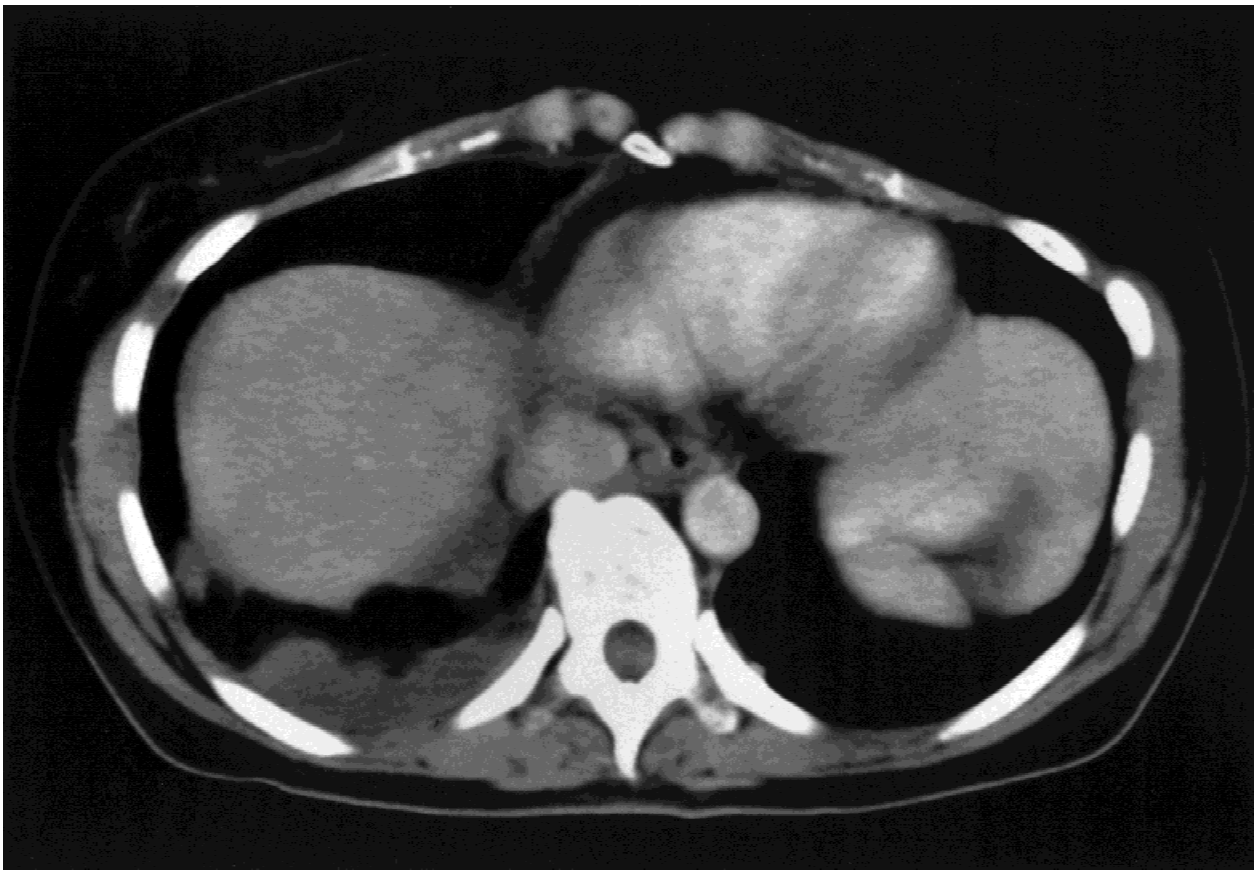

Fig. 2. CT showing right pleural effusion consistent with globular accumulation of mucinous tumor.

Despite extensive intraabdominal tumor accumulation, the extra abdominal spread of pseudomyxoma peritonei is a rare event. There are only a few reports of direct pleural involvement in the literature. Previously, reports related extension to the pleural space to be caused by direct tumor invasion, by surgical penetration of the hemidiaphragm, or by migration of tumor through the diaphragm [5,6,8].

In our case report, mucoid material accumulation in the pleural space was the result of flow of intraabdominal mucus through a congenital pleuroperitoneal communication. Defects in the diaphragm have been identified as a potential cause of pleural effusion in a patient with intraperitoneal fluid accumulation. Such pleuroperitoneal communications have been documented in patients with ascites secondary to hepatic cirrhosis and in individuals undergoing peritoneal dialysis [9,10]. Passage of ascitic fluid to the pleural space can also be attributed to leakage through lymphatic channels in the diaphragm. Technetium-99m and iodine-131 labeled albumin and technetium-99m labeled sulfur colloid have been used as intraperitoneal tracers to demonstrate the presence of pleuroperitoneal communications in cirrhotic patients and in renal failure patients undergoing peritoneal dialysis [10]. Activity appears in the pleural space within minutes of injection when a diaphragmatic defect is present, as opposed to several hours when tracers transit through lymphatic channels. In favor of the diagnosis of an occult communication is the unilaterality of the process.

A third cause of pleural effusion in a cancer patient is tumor invasion of the diaphragm. Tumor cells accumulate on peritoneal surfaces which absorb peritoneal fluid, such as the greater omentum and the undersurface of the diaphragm. Therefore, it is very common that patients with extensive peritoneal involvement will also present a thick layer of tumor on the undersurface of the diaphragm. Improvement in surgical techniques and the development of subphrenic peritonectomy procedures have enabled surgeons to achieve a complete cytoreduction of disease at this anatomic site. However, in selected cases, this may change the history of the disease. Violation of the diaphragm barrier either by stripping of its parietal peritoneum or by excising its tendinous portion allows the passage of tumor cells into the pleural cavity. In reviewing our data, 23 of 426 patients with pseudomyxoma peritonei who underwent cytoreductive surgery at our institution subsequently developed recurrence within the pleural space [11].

Pleural extension of mucinous tumor represents the development of pleural disease in patients in whom the

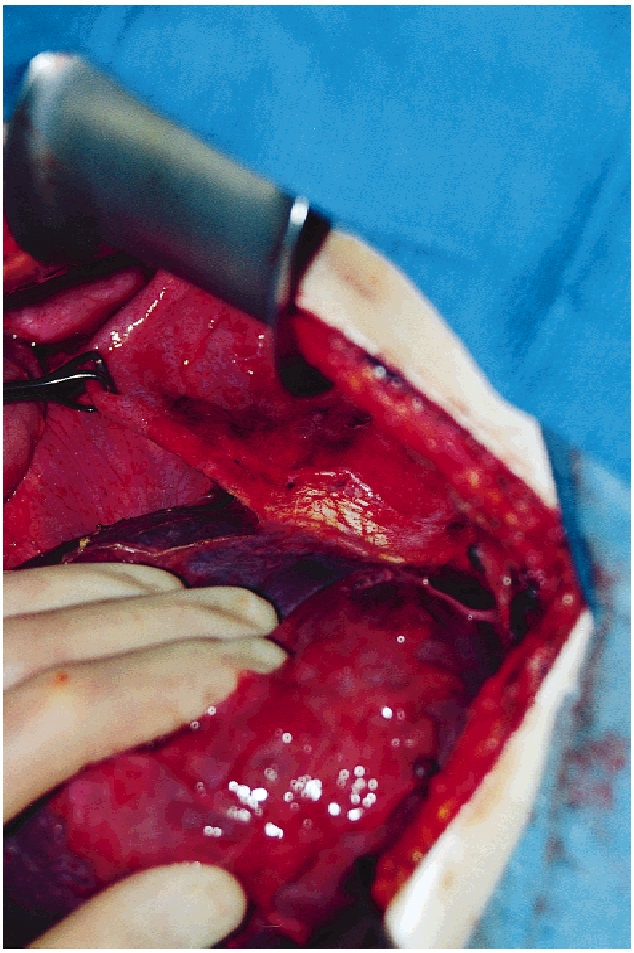

Fig. 3. As tumor was debulked from the undersurface of the right hemidiaphragm, three small circular defects in the central tendon were visualized. The mucinous tumor on Glisson's capsule is obvious.

diaphragm barrier is congenitally defective or has been iatrogenically compromised during abdominal cytoreduction. In both instances, tumor cells present on the peritoneum are disseminated into the pleural cavity. It is an important observation requiring adjustment of therapy for future patients. Using the same rationale as heated intraoperative intraperitoneal chemotherapy directed toward microscopic residual disease, heated intraoperative intrapleural chemotherapy may be of great interest to eradicate tumor cells that may have contaminated the pleural cavity. Intrapleural chemotherapy has been safely and successfully used in cases of pleuropulmonary diseases [12]. In our opinion, the timely treatment of pleural dissemination from pseudomyxoma peritonei depends largely on the ability to infuse intraoperative chemotherapy in the pleural space whenever the diaphragm has been penetrated. After the abdominal cytoreduction is completed, a Tenckhoff catheter is positioned for infusion and four suction drains for drainage. The abdomen is manually lavaged for 90 min with infusion and drainage of heated intraoperative intraperitoneal chemotherapy using mitomycin C (12.5 mg/m^2 for males, 10 mg/m^2 for females) in 3 l of 1.5% dextrose peritoneal dialysis solution. The temperature of the chemotherapy is progressively increased to reach an optimal temperature of 43°C. By enlarging the defect in the diaphragm, infusion of chemotherapy in the pleural space can be achieved during the same time period as the intraperitoneal chemotherapy. The entire pleural surface can be uniformly exposed to chemotherapy, therefore preventing tumor cell progression.

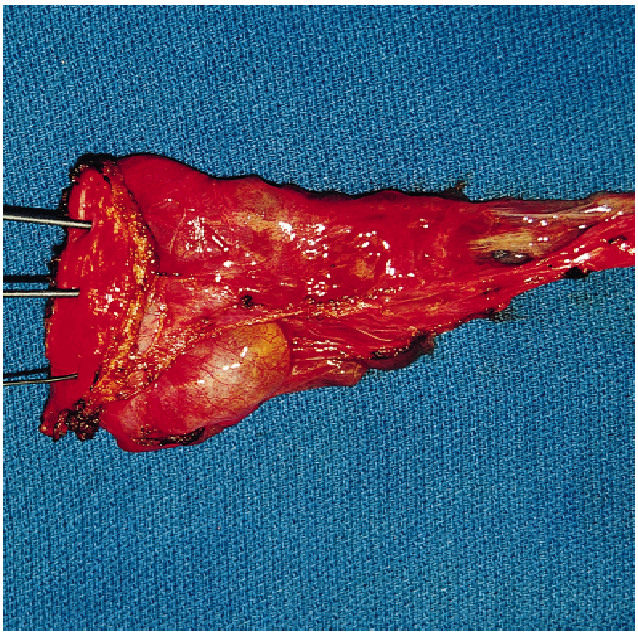

Fig. 4. Three defects in the tendinous central portion of the hemidiaphragm associated with mucoid material present above the right hemidiaphragm. The defects in the diaphragm were patent to blunt metal probes.

The present case demonstrates that a pleuroperitoneal communication should be considered in a patient with intraabdominal malignancy and a unilateral pleural effusion. Careful abdominal exploration may reveal diaphragmatic fenestration as the source of pleural tumor dissemination. In order to treat pleural tumor progression in such a situation or in cases of intraoperative diaphragm penetration, aggressive measures need to be taken. Exploration of the pleural cavity combined with heated intraoperative intraperitoneal and intrapleural chemotherapy is safe and should be considered.

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